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Bilateral Abductor Palsy in Neonates

Bigyan Raj Gyawali

Abstract

Paediatric airway compared to the adult has a considerable anatomical and physiological difference. Airway pathologies in neonates are very challenging in terms of diagnosis and management. Bilateral abductor paralysis is one of such situations. Despite being the second most common cause of stridor in paediatric population, the disease is still rare. Having a huge range of aetiological variations on one hand and the grievousness of pathology on the other, management of the disease is very challenging at times. Here, we present a review on various aetiological factors along with management of the disease.

Keywords: paediatric, airway, vocal fold, stridor, paralysis

1. Introduction

Airway pathologies in neonates are very challenging. Considering a wide variation in the anatomy and physiology compared to the adults, clinical approach to this population is entirely different.

Bilateral abductor paralysis means the failure of vocal cords to abduct resulting from denervation of recurrent laryngeal branch of vagus nerve. Although being a second most common cause of stridor in paediatric population, it is still a rare entity. The incidence in neonates is estimated to be 0.75 cases per million births per year [1]. There is no definite sex predilection. With no single definable cause and being a common presentation of wide range of pathologies, for example, congenital, neurological, traumatic etc., this is a topic that has to be critically explored.

2. Anatomy

2.1 Laryngeal framework

Paediatric airway differs considerably from adult airway. Location of the larynx in the neonates is high up in the neck opposite to cervical third and fourth vertebra which gradually descends down with age. Thyrohyoid membrane is short compared to the adult; thus, the thyroid notch lies either posterior or just inferior to the hyoid bone. Subglottis is the narrowest part of the larynx in neonates and the abundance of soft tissue in the subglottis and supraglottis make them vulnerable to swelling during inflammation. In the glottis, arytenoids make up the 50% of antero-posterior length and the vocal cords, next 50%.

2.2 Laryngeal musculature

Larynx comprises of two groups of muscles: intrinsic and extrinsic. Intrinsic muscles are usually responsible for various movements of vocal cords. Posterior cricoarytenoid is the only muscle of the larynx that abducts the vocal cords. Lateral cricoarytenoid and interarytenoid muscles act as adductor of the vocal cord. Thyroarytenoid and cricothyroid muscles act as tensor of the vocal cords.

2.3 Nerve supply

Both sensory and motor supplies to larynx come from vagus nerve via superior and recurrent laryngeal nerve. All the muscles of the larynx are supplied by recurrent laryngeal nerve except for the cricothyroid, which is supplied by the external branch of superior laryngeal nerve. Sensory supply of the supraglottis and glottis is provided by the internal branch of superior laryngeal nerve, and the subglottis is provided by recurrent laryngeal nerve.

The abductor fibers in the recurrent laryngeal nerve are phylogenetically newer compared to the adductor fibers and occupy the periphery of the nerve. This makes them more vulnerable to injury from any organic lesion (*Semon's law*).

3. Aetiology

3.1 Congenital causes

Congenital bilateral abductor palsy may present immediately or within few days of birth. Child may present with stridor, weak cry, cyanosis, or difficulty in feeding. Aetiological factors can be broadly classified into genetic or nongenetic.

3.1.1 Genetic

Bilateral abductor palsy presenting with the genetic etiology is very rare. Presentation may be with an isolated paralysis of vocal folds or as a spectrum of various clinical features. Plott syndrome, described by Plott in 1964 [2], is a rare entity demonstrating X-linked recessive inheritance. Patients usually present with bilateral vocal fold paralysis and mental retardation. Autosomal dominant mode of inheritance has also been reported [3–5]. Other syndromic associations may be Down's syndrome, Moebius syndrome, congenital myasthenic syndrome, 22q deletion syndrome and Goldenhar syndrome [6]. Rarely, it may be an isolated presentation of brainstem discontinuity, for example, medullary defect. This is a very rare congenital abnormality where there is a segmental discontinuity of brainstem [7].

3.1.2 Nongenetic

Several developmental anomalies of brain and brainstem may secondarily present with bilateral abductor palsy. Arnold Chiari malformation, congenital hydrocephalus, meningomyelocele, syringomyelia, syringobulbia, etc. are such examples.

3.2 Acquired causes

Various etiological factors have been mentioned in the literatures so far. Following are the common acquired causes for bilateral abductor palsy.

3.2.1 Idiopathic

In majority of cases with bilateral vocal fold palsy, no aetiological factors can be found. However, 50% these patients tend to recover completely or partially within a period of 1–2 years. The improvement is thought to be a result of delayed maturation of vagal nuclei [8].

3.2.2 Traumatic

Trauma either to the vocal cords and cricoarytenoid joint or to the vagus nerve itself can present with bilateral abductor palsy in neonates. Commonest causes are birth trauma from instrumental delivery, cardiac surgeries, mediastinal surgeries, for example, PDA ligation, etc.

3.2.3 Perinatal encephalopathy

This is another recognized cause for bilateral abductor palsy in neonatal period. It can result from various causes like perinatal hypoxia or ischaemia, birth trauma or infection.

3.2.4 Neurological disorders

Various neurological disorders like Myasthenia Gravis, Charcot-Marie tooth Disease and multiple sclerosis can present with bilateral vocal cord paralysis; however, there are features of other neurological dysfunctions as well. These conditions to present neonatally are also exceedingly rare.

3.2.5 Inflammatory

Neuritis of recurrent laryngeal nerve resulting from bacterial or viral infections can cause bilateral abductor palsy. In the pre-antibiotic era, Syphilis was considered one of the common causes, which now has become very rare. Viruses such as Influenza and Herpes Simplex virus are also reported of causing bilateral abductor palsy [9].

4. Clinical evaluation

A detailed history and a thorough clinical examination will most of the times lead to the inciting cause. Clinical history should focus on any significant perinatal events like maternal infection, maternal drug use, time and mode of delivery, instrumentation during labour, birth weight and neonatal infection. A family history should be taken as there are reported cases of this entity being inherited within the family members.

A complete head to toe examination of the affected child should always be done as there may be signs that can guide towards the aetiological cause. Neurological examination should include motor and sensory evaluation along with examination of cranial nerves specially 9th, 10th and 11th which exit commonly through the jugular foramen. Features of raised ICP such as bulged fontanelle, papilloedema should be sought. Chest and heart should be examined thoroughly. ENT examination should focus on palatal mobility, tongue mobility and presence of gag reflex.

The child may have a wide range of presentations. Most of the cases are asymptomatic until they present with sudden onset stridor preceded by upper respiratory

tract infections. Other cases might present with stridor immediately following birth or few days after birth. Stridor is usually during the inspiratory phase and tends to worsen on crying where it is not unusual for a child to become cyanotic. Since the vocal folds are in adducted position change in voice is usually not encountered. Other clinical features may be difficulty in feeding, recurrent pneumonia, O₂ dependence and repeated failure to extubate. Although nonspecific, these clinical features always warrant a laryngeal evaluation.

5. Investigations

5.1 Endoscopic evaluation

An in-office flexible laryngoscopy can be done as an initial diagnostic procedure to evaluate the mobility of bilateral vocal folds, adequacy of glottic chink and to rule out other associated laryngeal anomalies such as laryngomalacia, laryngeal cleft, anterior or posterior glottic stenosis, etc. In addition, it is also important to look for velopharyngeal closure. Functional endoscopic evaluation of swallowing should be done in those who have recurrent aspiration. A rigid endoscopic evaluation under intravenous anaesthesia will also be required to assess the subglottis, trachea and bronchi as it is not uncommon to encounter second laryngeal pathology such as subglottic stenosis and tracheobronchomalacia. Cricothyroid joint should also be assessed for its mobility and to rule out any traumatic dislocation.

5.2 Imaging

5.2.1 Ultrasonography

Ultrasound can detect the neurological abnormalities like meningocele, Arnold Chiari malformation, hydrocephalus prenatally which can have associated bilateral abductor palsy. In neonates with significant morbidity, laryngeal ultrasound can also be done to detect vocal fold palsy and any associated pathologies in the neck [10].

5.2.2 Magnetic resonance imaging

MRI scan of brain, brainstem, neck and mediastinum should be done routinely as pathologies like Arnold Chiari malformation, intraventricular hemorrhage, meningocele, brainstem dysgenesis, neck or mediastinal masses can be identified easily.

5.2.3 X-ray

Chest x-ray can give the evidence of aspiration if present as well as any associated cardiac, lungs or mediastinal pathologies.

5.3 Serology

Cases where there are features of viral infection, viral serology should always be done specially for Herpes Simplex and Influenza virus. Anti-viral drugs if started early have good outcome in these cases. Also, serological tests for syphilis may be warranted if there are features of congenital syphilis.

5.4 Laryngeal electromyography

It is usually done to differentiate between vocal cord immobility due to denervation and mechanical fixation, for example, cricoarytenoid joint fixation and posterior glottis stenosis.

6. Management

Primary pathology should always be sought and managed accordingly as in most of the cases correcting the primary pathology reverts the mobility of vocal folds as well, for example in Arnold-Chiari malformation a shunt procedure may lower the intracranial pressure and thus reduce the stretching of the vagus nerve. In idiopathic cases, where no obvious inciting pathology could be found a watchful waiting up to 24 months can be done. Many of these cases gain normal or partial mobility of the cords during this period.

Most of the neonates with bilateral abductor palsy are asymptomatic until they present with an acute respiratory compromise due to upper respiratory tract infection. Management can be broadly discussed under following two headings.

6.1 Medical

It has a very limited role in the management. Corticosteroids can be given in cases presenting with stridor to reduce vocal cord oedema. Antivirals, for example, Acyclovir, have shown to hasten the recovery in cases with suspected viral aetiology.

6.2 Surgical management

There has been a huge advancement in the field of airway surgery in the past century with various surgical modalities coming forth. Here, we discuss various surgical treatment modalities for bilateral vocal cord palsy.

6.2.1 Tracheostomy

Tracheostomy may not always be required. However, in cases presenting with stridor immediate establishment of an alternate airway by tracheostomy is the earliest and safest procedure that can be performed. Tracheostomy provides a secured airway following which a specific management approach can be planned. Following tracheostomy, the tracheostomy tube care also poses a challenge as it is not uncommon for neonatal tracheostomy tube to get easily blocked with tracheal secretions. It is an ideal approach to wait until the child is 2 years old before embarking on other surgical procedures as it will allow a time period for spontaneous recovery.

6.2.2 Lateralization of vocal cord

This is a temporary procedure where the vocal cords are lateralized with a suture. There is a lesser risk of aspiration and this procedure can be reverted once the vocal cord function is back. One of the drawbacks of this procedure is that the suture may give away and the cord may be medialised again. It is reserved for the cases where recovery of vocal cord function is anticipated.

6.2.3 Posterior cordotomy

This is an irreversible procedure. Endoscopic LASER cordotomy was popularized by Dennis and Kashima [11]. In this procedure, a transection is carried out through true vocal fold, ventricle and false vocal fold from the vocal process creating a wedge shaped defect in the posterior glottis. The drawbacks of this surgery are chances of aspiration and change in voice.

6.2.4 Arytenoidectomy

Similar to posterior cordotomy, this is also an irreversible procedure aimed at creating a defect in posterior glottis. However, in this surgery most of the arytenoid is removed. This procedure also carries a risk of change in voice and aspiration. This procedure can be carried out via both open and endoscopic approach.

6.2.5 Combined posterior cordotomy and arytenoidectomy

6.2.6 Posterior cricoid split and grafting

This surgery aims at widening the glottic aperture by expanding the posterior laryngeal commissure with the help of costal cartilage. It can be performed via both open and endoscopic approach. The advantage of this procedure is that integrity of the vocal folds and arytenoids is kept intact and there are also minimal chances of aspiration.

6.2.7 Reinnervation

Reinnervation using ansa cervicalis nerve-muscle pedicle transfer or phrenic nerve transfer to posterior cricoarytenoid muscle can be done. The results are, however, not consistent and more clinical human trials are further needed to draw a definite conclusion.

7. Conclusion

Bilateral abductor palsy in neonates, although the second most common cause for stridor in paediatric population, is still a rare and clinically challenging entity. It can be a presentation of various pathologies, both genetic and non-genetic. Investigations should be guided by the mode of presentation and the possible aetiological factors, however, MRI brain should be done in all cases to rule out anomalies such as Arnold-Chiari malformation. Watchful waiting is always a standard approach until the child is 2 years old in idiopathic cases as significant number of cases tend to improve by 1–2 years.

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Author details

Bigyan Raj Gyawali

Institute of medicine, Tribhuvan University Teaching hospital, Kathmandu, Nepal

*Address all correspondence to: bigyan.gyawali@gmail.com

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